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Cullen's sign – Case report with a review of the literature

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ABSTRACT

INTRODUCTION: Cullen's sign is described as superficial oedema with bruising in the subcutaneous fatty tissue around the peri-umbilical region. It is also known as peri-umbilical ecchymosis.
PRESENTATION OF CASE: We report a case of a fifty-three year old gentleman who presented with non-specific abdominal pain. Significant bruising was present within the subcutaneous fatty tissue on the anterior abdominal wall in keeping with 'Cullen's sign.' Of note he denied any alcohol intake and his amylase was normal. A diagnosis of pancreatitis was made following a CT scan of his abdomen.
DISCUSSION: A search of the entire English literature using PubMed with the phrase 'Cullen's sign' has been performed. Papers were reviewed in relation to recognition of this clinical sign, differential diagnosis, and management.
CONCLUSION: Our case and review of the literature highlights the rarity of this clinical sign which a clinician may encounter. In addition we draw to attention the importance of having knowledge of the underlying possible conditions which may lead to this sign, and can be vital in successful acute management.

1. Case report

A fifty three year old gentleman presented to the emergency department with acute epigastric and central abdominal pain. The pain radiated to his back and was constant in nature. He described the pain as being at its peak intensity three days prior to admission. His pain was associated with nausea and vomiting. His bowels were opening normal and there were no urinary symptoms. He denied any chest pain. His past medical history included a long history of chronic back pain. He had no surgical history of note. He smoked forty roll up tobacco cigarettes per day, and denied any alcohol intake. His medication included naproxen and there were no allergies of note.

Clinical observations revealed a blood pressure of 140/88 mm Hg with pulse rate of 90 beats per minute. His respiratory rate was 18 breaths per minute and oxygen saturation was 97% on room air. He was alert with a Glasgow Coma Score of 15/15. Examination revealed no evidence of jaundice, anaemia or lymphadenopathy. Cardiovascular and respiratory examinations were essentially normal. Abdominal examination revealed significant bruising primarily across the upper abdomen (Fig. 1) and to a lesser degree in the supra pubic region. Palpation elicited tenderness in upper abdominal region. There was no guarding or rebound tenderness, but evidence of peritonism with regards to the bruising. Per rectal examination revealed soft faeces. Noting the bruising on the anterior abdominal wall, the patient was questioned but denied any history of trauma. The bruising in the subcutaneous fatty tissue was in keeping with 'Cullen's sign' and hence a clinical impression of acute pancreatitis was made. There was no bruising present to the lateral abdominal wall or back and hence Grey Turner's sign was not present.

He continued to remain haemodynamically stable. Blood tests revealed haemoglobin (Hb) of 14.1 gm/dL, with an elevated mean corpuscular volume (MCV) of 104.6 fl. Urea and electrolytes (U&Es), liver function tests (LFTs) and clotting were within normal range. His c-reactive protein (CRP) was 3 mg/L and interesting his amylase returned a normal result of 72 IU/L (normal range 25–125). His lactate was 1.5 mmol/L (normal range 0.5–2.2) and arterial blood gases performed also revealed parameters within normal range. A chest X-ray (Fig. 2) revealed normal heart and mediastinal contours with the lungs clear. There was no free subdiaphragmatic gas identified. Abdominal X-ray (Fig. 3) performed revealed no acute features of note.

His initial management included appropriate analgesia with anti-emetic and the commencement of intravenous fluids. An ultrasound (Fig. 4) of the abdomen revealed the liver parenchyma to be diffusely hyper echoic in keeping with fatty infiltration. No gallstones were identified and no intrahepatic biliary dilatation was seen. The pancreas was not visualised adequately due to overlying gas and hence a computed tomographic scan (CT) was recommended. CT imaging revealed normal enhancement of the entire pancreatic gland but with change in keeping with non-complicated acute pancreatitis. There were no fluid collections or necrosis (Fig. 5).

On further questioning the patient admitted that he was a chronic alcoholic, and usually consumed four to five cans of strong cider per day. This explained the examination finding of Cullen's

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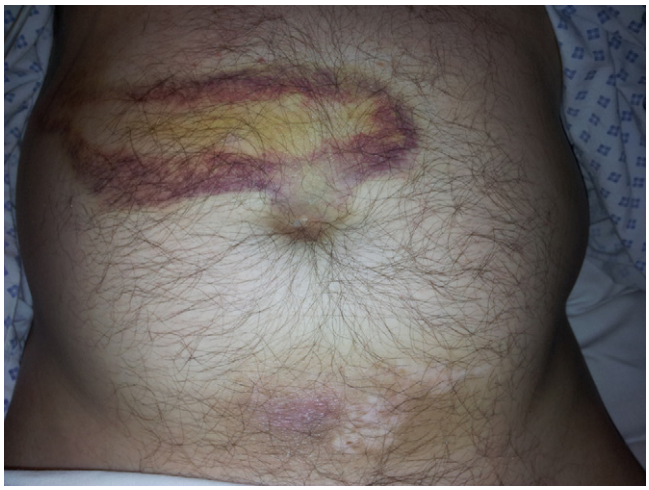

Fig. 1. Cullen's sign.

sign, the elevated MCV, normal amylase results and CT imaging. A diagnosis of delayed presentation of ethanol induced acute on chronic pancreatitis was hence made.

He remained haemodynamically stable whilst being treated for alcohol withdrawal medication. He made steady progress and was discharged home after six days. He was followed up in outpatient clinic at six weeks and had returned to his normal activities with no on-going problems. There have been no further admissions to date, at six months since discharge.

2. Discussion

A search of the entire English literature using PubMed with the phrase 'Cullen's sign' resulted in 43 articles. Articles were dated between 1937 and 2008. There were 11 papers which were deemed relevant in relation to the presence of the clinical sign, differential diagnosis and management.

Our case and review highlights the rarity of this clinical sign in keeping with the literature which a clinician may be confronted with. In addition we draw to attention the importance of having knowledge of the underlying possible conditions, which can be vital in successful acute management.

Fig. 2. CXR.

Fig. 3. AXR.

Cullen's sign is described as superficial oedema with bruising in the subcutaneous fatty tissue around the peri-umbilical region. This is also known as peri-umbilical ecchymosis. It is most often recognised as a result of haemorrhagic pancreatitis. The sign was named after Thomas S. Cullen (1868–1953), a Canadian Gynaecologist who researched gynaecological disease including uterine cancer and ectopic pregnancy, and became a Professor at John Hopkins Hospital. He first described the sign in 1918 after a case of a ruptured extra uterine pregnancy.[1]

The sign can take 2–3 days before appearance, and may be used as a clinical sign to help the diagnosis of acute pancreatitis. Cullen's sign may co-exist with Grey Turner's sign (bruising in the

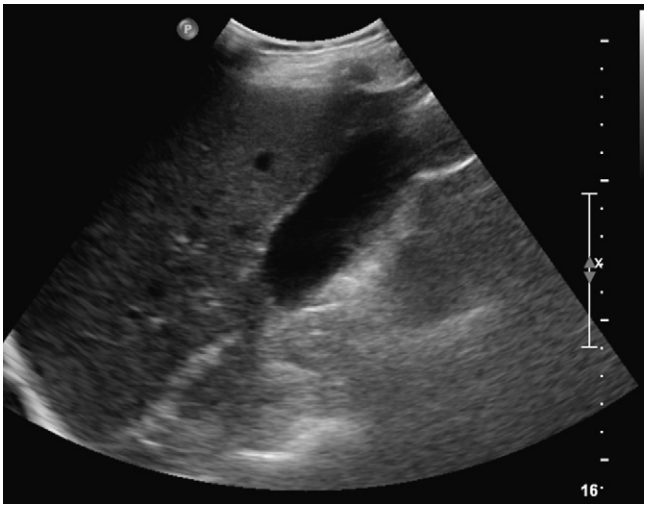

Fig. 4. US.

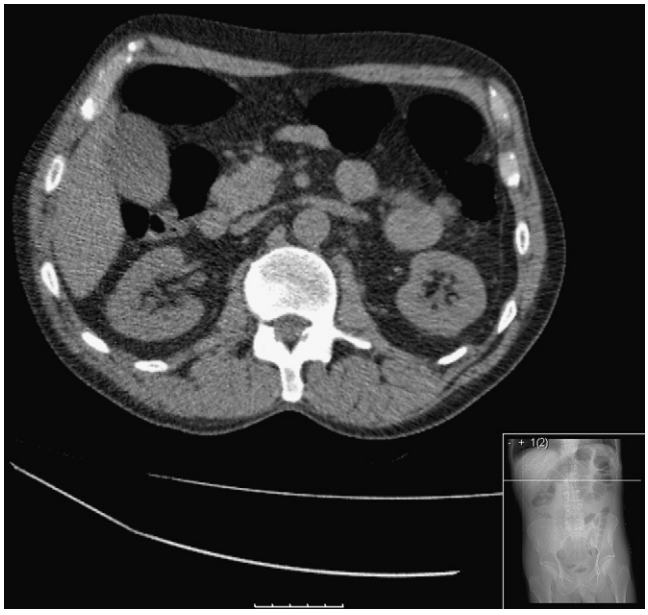

Fig. 5. CT.

lateral aspect of the abdominal wall or the flanks.[2,3] It is thought that one or both of the signs may be present in 1–3% of patients with acute pancreatitis.[3–6] Either of these signs can be an indication of pancreatic necrosis with retroperitoneal or intra-abdominal bleeding.

The discoloration around the peri-umbilical region can vary in colour from green/yellow to purple, but varies according to the stage of breakdown of the red blood cells. Pancreatic enzymes have been suggested as a mechanism leading to the discoloration of the abdominal wall adipose tissue, but this has not been widely accepted due to the presence of peri-umbilical ecchymosis presenting in the absence of pancreatitis.[4,7]

Cullen's sign occurs due to tracking of haemorrhagic fluid from the retro-peritoneum along the gastrohepatic and falciform ligament to the umbilicus.[3] In contrast Grey Turner's sign is due to the diffusion of blood from the posterior pararenal space to the lateral edge of the quadratus lumborum muscle, where a defect in the transversalis fascia permits access to the abdominal wall musculature and subsequently tissue of the flank.[3,6]

A wide range of causes of Cullen's sign have been documented in the literature, and these have been highlighted in Table 1.[7–10]

Table 1
Causes of Cullen's sign in the medical literature.

- Pancreatitis
- Ruptured ectopic pregnancy
- Perforated duodenal ulcer
- Percutaneous liver biopsy
- Ruptured abdominal aortic aneurysm
- Metastatic thyroid cancer
- Post angiographic/radiological intervention
- Pancreatic/abdominal trauma
- Amoebic liver abscess
- Ovarian enlargement secondary to primary hypothyroidism
- Metastatic oesophageal cancer
- Ruptured spleen
- Ruptured common bile duct
- Hepatocellular carcinoma
- Hepatic lymphoma
- Ruptured internal iliac artery aneurysm

Differential diagnosis can include peri-umbilical cellulitis, Sister Mary Joseph's sign or metastatic spread of an intra-abdominal malignancy to the umbilicus, subcutaneous administration of heparin, psoriasis, endometriosis with umbilical involvement, ulceration of a re-canalised umbilical vein in the setting of cirrhosis, adenocarcinoma of urachal remnants and squamous cell carcinoma.[7]

Cullen's sign should alert the clinician to the possibility of significant intra-abdominal bleeding or pathology and hence further investigation and management should be commenced expediently.

Mortality rate of 37% has been reported in patients with acute pancreatitis with the presence of Cullen's or Grey Turner's sign, and hence recognition of the clinical sign is imperative in initiating management with urgency.[4] This is especially crucial as with the development of multiple diagnostic modalities the recognition and importance of the sign may be on the decline.

Fujiwara et al. reviewed the CT scan findings in 277 patients with acute pancreatitis.[11] Inflammatory changes involving the anterior abdominal wall were present in 5 of 277 cases (1.8%). All five patients recovered well and the authors concluded that Cullen's sign or anterior extension of acute pancreatitis does not directly mean extensive retroperitoneal involvement. The results of this study are in keeping with our case, but as highlighted earlier, knowledge of causes other than pancreatitis leading to Cullen's sign may be critical.

Conflict of interests

The authors declare that they have no competing interests.

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None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author's contribution

GR and MU were actively involved in the clinical care of the patient. GR and MU performed the literature review. GR, MU, NY and GT drafted the paper, acquired the figures and revised the manuscript. All authors have read and approved the final manuscript.

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